

# Calcifying Cystic Odontogenic Tumor

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## History

A 38 year old male presented with an 18 month history of an asymptomatic swelling in the anterior mandibular vestibule. The clinical exam revealed a firm and expansile lesion involving the buccal cortical plate. The lesion extended from tooth #23 to tooth # 27. No clinical evidence of caries or periodontal disease was detected and the patient denied history of trauma to the area. The radiographic exam revealed a lesion in the anterior mandible. Vitality tests were within normal limits.

## Radiographic Features

Imaging studies revealed a well defined unilocular radiolucency extending from tooth #23 to tooth # 27 (Fig. 1). Within the center of the radiolucency and extending inferiorly, indistinct opacities were noted. The panoramic film showed what appeared to be root resorption of teeth #23–#26; however review of periapical films ruled resorption out (Fig. 2).

## Treatment

The patient was referred to an oral surgeon for a biopsy and treatment. The lesion was enucleated and submitted for histologic evaluation.

## Diagnosis

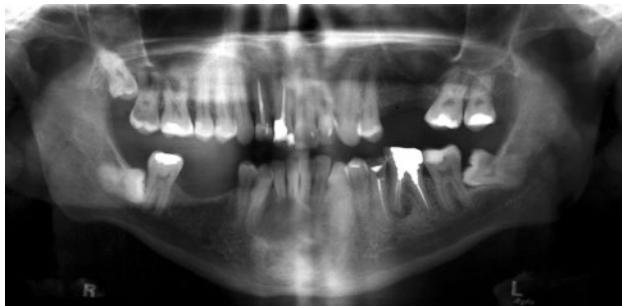
The microscopic examination revealed a cystic lesion with a fibrous capsule lined by an outer layer of odontogenic epithelium four to six cells in thickness. The basal cells of the outer layer were columnar resembling ameloblasts. A layer composed of loosely arranged epithelium resembling stellate reticulum was noted overlying the first layer (Fig. 3). Numerous ghost cells and calcifications were identified within the epithelial lining (Fig. 4).

## Discussion

The calcifying cystic odontogenic tumor (CCOT), also known as calcifying odontogenic cyst (COC) or Gorlin cyst is a rare developmental lesion which arises from odontogenic epithelium. Although the lesion has been commonly recognized as a benign odontogenic cyst since Gorlin et al. first described it in 1962 [1], this pathologic entity encompasses a spectrum of clinical behavior and histopathological features including cystic, solid (neoplastic) and aggressive (malignant) variants [2–7]. As a result of this diversity, different classification schemes and nomenclature for the lesion and its variants have been suggested. According to Hong et al., although most pathologists favor the term calcifying odontogenic cyst (which emphasizes the cystic nature, odontogenic origin, and calcifying

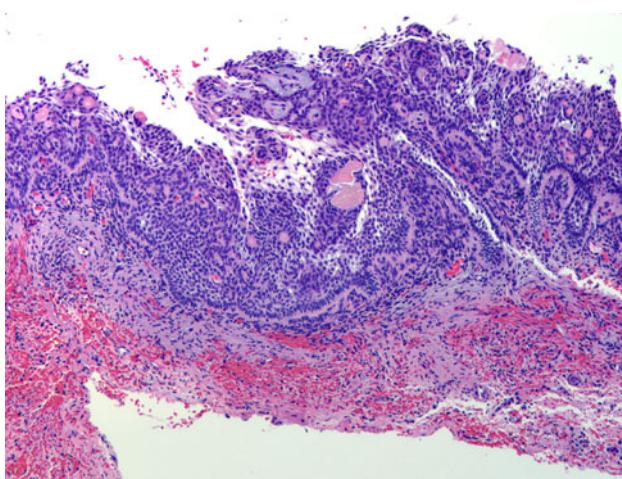
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**Fig. 1** Panoramic radiograph shows a unilocular radiolucency in the anterior mandible

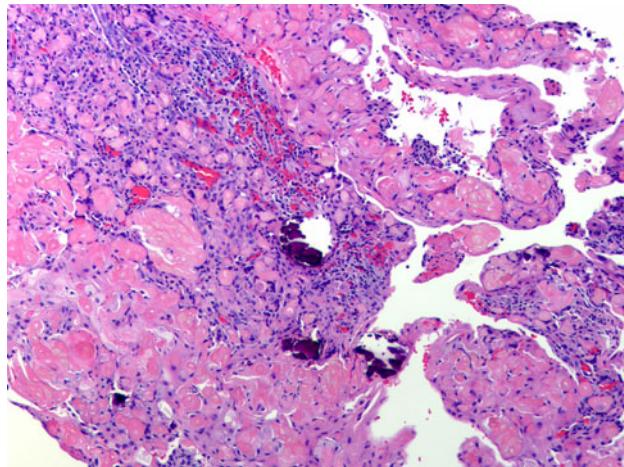
**Fig. 2** Periapical radiograph reveals unilocular radiolucency with lack of root resorption



**Fig. 3** Photomicrograph showing cyst lining composed of an outer layer of columnar basaloid epithelium and an inner layer resembling stellate reticulum of enamel organ

potential of these lesions); others consider the lesion to be a neoplasm [5]. The *World Health Organization Classification of Head and Neck Tumors* classifies the lesion as an odontogenic tumor and names it calcifying cystic odontogenic tumor [8].

The calcifying cystic odontogenic tumor can occur in any location of the oral cavity and approximately 65–67.5% of cases occur in the anterior jaws [4, 7]. CCOTs occur with equal frequency in the maxilla and mandible



**Fig. 4** Photomicrograph showing numerous ghost cells and calcifications

[2–4, 8, 9] and demonstrate no gender predilection [1–3, 8, 9]. The lesion occurs in a broad age group with a peak incidence in the second decade of life [2, 7, 9–11]. The CCOT is believed to arise from odontogenic epithelial remnants trapped within the bones of the maxilla and mandible or gingival tissues [11], therefore CCOTs can develop either centrally (intraosseous) or peripherally (extraosseous) [2, 3, 8, 9]. The majority of cases (86–98%) demonstrate a cystic architecture while the solid (neoplastic) form comprises 2–16% of cases [4].

Most calcifying cystic odontogenic tumors are asymptomatic, often discovered incidentally on radiographic exams. Because the lesion arises in tooth bearing areas of the jaws or gingiva, CCOTs are often located in a periapical or lateral periodontal relationship to adjacent teeth [3]. Radiographically, CCOTs are well-delineated and appear as a unilocular or multilocular radiolucency [2–4, 6, 8–10] with calcifications of variable density noted in one-third to one-half of cases [4]. CCOTs can occur alone or in association with other odontogenic tumors such as odontomas (20%), adenomatoid odontogenic tumors and ameloblastomas [4]. Root resorption and divergence are common radiographic findings [4, 8] and an association with an impacted tooth occurs in approximately one-third of cases [4, 9]. Asymptomatic swelling is a common presenting sign in both extraosseous and intraosseous locations with expansion of the buccal and/or lingual cortical plates often occurring with the latter [2, 3, 8, 9].

Histologic features include a cyst lining composed of an outer layer of a columnar basaloid odontogenic epithelium and an inner layer resembling stellate reticulum of the enamel organ. Characteristic features for CCOT include the presence of ghost cells and/or calcifications within the cyst lining or fibrous capsule.

Enucleation is the treatment of choice for most intraosseous CCOTs with few recurrences reported in the literature [8]. The extraosseous form is treated with surgical excision and recurrences for this type have not been reported [4]. The prognosis for both intraosseous and extraosseous CCOTs is good.

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